What is claimed is:

1. A transgenic nonhuman mammal having a transgene comprising:
a promoter and enhancer from the same mammary gland specific gene;
a secretory DNA segment encoding a signal peptide functional in mammary secretory cells of the transgenic nonhuman mammal, and
a recombinant DNA segment encoding acid .alpha.-glucosidase operably linked to the secretory DNA segment to form a secretory-recombinant DNA segment, the secretory-recombinant DNA segment being operably linked to the promoter and enhancer, and wherein the secretory DNA segment is an acid a glucosidase secretory DNA segment or is from the same mammary-gland specific gene as the promoter and enhancer;
wherein the transgene, in an adult form of the nonhuman mammal or a female descendant of the nonhuman mammal, expresses the secretory-recombinant DNA segment in the mammary secretory cells to produce acid .alpha.-glucosidase that is processed and secreted by the mammary secretory cells into milk in a recoverable amount with .alpha.-glucosidase catalytic activity.
2. The transgenic nonhuman mammal of claim 1, wherein the concentration of the acid .alpha.-glucosidase in the milk is at least 100 .mu.g/ml.
3. The nonhuman transgenic mammal of claim 1, wherein the secretory DNA segment is an acid .alpha.-glucosidase secretory DNA segment.
4. The transgenic nonhuman mammal of claim 1, wherein the human acid .alpha.-glucosidase is secreted into milk in a form that can be taken up by muscle cells.
5. The nonhuman transgenic mammal of claim 1, wherein the acid .alpha.-glucosidase is human.
6. The nonhuman transgenic mammal of claim 5, that is a mouse or rabbit.
7. The nonhuman transgenic mammal of claim 6, wherein the recombinant DNA segment is cDNA.

8. The nonhuman transgenic mammal of claim 6, wherein the recombinant DNA segment is genomic.

9. The nonhuman transgenic mammal of claim 6, wherein the recombinant DNA segment is a cDNA-genomic-DNA hybrid.

10. A method for producing acid .alpha.-glucosidase, the method comprising: recovering milk from the adult form of the transgenic nonhuman mammal of claim 1 or its female descendant, wherein said milk contains a recoverable amount of acid .alpha.-glucosidase with catalytic activity.

11. The method of claim 10, further comprising incorporating the milk into a food product.

12. The method of claim 10, further comprising purifying the acid .alpha.-glucosidase from the milk.

13. The method of claim 12, wherein the acid .alpha.-glucosidase is purified to at least 95% pure from other components of the milk.

14. The method of claim 13, further comprising mixing the acid .alpha.-glucosidase with a pharmaceutical carrier for intravenous, intradermal, intramuscular or oral administration.

15. Milk from the transgenic nonhuman mammal of claim 1, the milk comprising human acid .alpha.-glucosidase in a recoverable amount.

16. The milk of claim 15, wherein the concentration of the human acid .alpha.-glucosidase is at least 100 .mu.g/ml.

17. A composition comprising human acid .alpha.-glucosidase with catalytic activity and capacity to be taken up by muscle cells in a patient and milk of the nonhuman transgenic mammal of claim 1.

18. A pharmaceutical composition for parenteral administration to a human patient comprising human acid .alpha.-glucosidase with catalytic activity and in a therapeutically effective dosage to treat a patient suffering from Pompe's disease; and a pharmaceutical carrier, the composition being free of other human proteins present in its natural environment.

19. The pharmaceutical composition of claim 18, wherein the pharmaceutical carrier is for intravenous administration.

20. The pharmaceutical composition of claim 18, wherein the human acid .alpha.-glucosidase is purified to homogeneity.

21. A method of treating a patient with Pompe's disease, comprising: administering to the patient a therapeutically effective amount of human acid alpha glucosidase.

22. The method of claim 21, wherein the patient is administered at least 10 mg/kg body weight per week.

23. The method of claim 21, wherein the patient is administered at least 15 mg/kg body weight per week.

24. The method of claim 21, wherein the patient is administered at least 20 mg/kg body weight per week.

25. The method of claim 21, wherein the patient is administered at least 30 mg/kg body weight per week.

26. The method of claim 21, wherein the patient is administered at least 45 mg/kg-60 mg/kg body weight per week.

27. The method of claim 21, wherein the patient is administered at least 60 mg/kg body weight per week.

28. The method of claim 21, wherein the patient is administered at least 120 mg/kg body weight per week.

sub a' 29. The method of any of claims 21-28, wherein the patient is administered a single dosage of alpha-glucosidase per week.

30. The method of any of claims 21-28, wherein the patient is administered two dosages of alpha-glucosidase per week.

31. The method of any of claim 21-28, wherein the patient is administered three dosages of alpha-glucosidase per week.

32. The method of any of claims 21-31, wherein the amount is administered per week for a period of at least four weeks.

33. The method of any of claims 21-31, wherein the amount is administered per week for a period of at least 24 weeks.

34. The method of any of claim 21-33, wherein the alpha-glucosidase is administered intravenously.

35. The method of claim 21, wherein the alpha-glucosidase was produced in milk of a transgenic mammal.

36. The method of claim 21, wherein the alpha-glucosidase was produced from a CHO cell-line.

37. The method of claim 21, wherein the patient has infantile Pompe's disease.

38. The method of claim 21, wherein the patient survives to be at least one year old.

39. The method of claim 21, wherein the patient has juvenile Pompe's disease.

40. The method of claim 21, wherein the patient has adult Pompe's disease.

41. The method of claim 21, wherein the alpha-glucosidase is predominantly in a 110 kD form.

42. The method of claim 21, further comprising monitoring a level of human acid alpha glucosidase in the patient.

43. The method of claim 21, further comprising administering a second dosage of human acid alpha glucosidase if the level of alpha-glucosidase falls below a threshold value in the patient.

44. The method of claim 21, wherein the human alpha glucosidase is administered intravenously and the rate of administration increases during the period of administration.

45. The method of claim 44, wherein the rate of administration increases by at least a factor of ten during the period of administration.

46. The method of claim 44, wherein the rate of administration increases by at least a factor of ten within a period of five hours.

47. The method of claim 21, wherein the patient is administered a series of at least four dosages, each dosage at a higher strength than the previous dosage.

48. The method of claim 47, wherein the dosages are a first dosage of 0.03-3 mg/kg/hr, a second dosage of 0.3-12 mg/kg/hr, a third dosage of 1-30 mg/kg/hr and a fourth dosage of 2-60 mg/kg/hr.

49. The method of claim 47, wherein the dosages are a first dosage of 0.11 mg/kg/hr, a second dosage of 1-4 mg/kg/hr, a third dosage of 3-10 mg/kg/hr and a fourth dosage of 6-20 mg/kg/hr.

50. The method of claim 47, wherein the dosages are a first dosage of 0.25-4 mg/kg/hr, a second dosage of 0.9-1.4 mg/kg/hr, a third dosage of 3.6-5.7 mg/kg/hr and a fourth dosage of 7.2-11.3 mg/kg/hr.

51. The method of claim 23, wherein the dosages are a first dosage of 0.3 mg/kg/hr, a second dosage of 1 mg/kg/hr, a third dosage of 4 mg/kg/hr and a fourth dosage of 12 mg/kg/hr

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52. The method of any of claims 47-51, wherein the first, second, third and fourth dosages are each administered for periods of 15 min to 8 hours.

53. The method of any of claims 47-51, wherein the first, second, third and fourth dosages are administered for periods of 1 hr, lhr, 0.5 hr and 3 hr respectively.

54. A pharmaceutical composition comprising human acid alpha . glucosidase, human serum albumin, and a sugar in a physiologically acceptable buffer in sterile form.

55. The pharmaceutical composition of claim 54 comprising human acid alpha glucosidase, human serum albumin, and glucose in sodium phosphate buffer.

56. A pharmaceutical composition comprising alpha glucosidase, mannitol and sucrose in an aqueous solution.

57. The pharmaceutical composition of claim 56, wherein the sugar comprises mannitol and sucrose and the concentration of mannitol is 1-3% w/w of the aqueous solution and the concentration of sucrose is 0.1 to 1 % w/w of the aqueous solution.

58. The pharmaceutical composition of claim 56, wherein the concentration of mannitol is 2% w/w and the concentration of sucrose is 0.5% w/w.

59. A lyophilized composition produced by lyophilizing a pharmaceutical composition comprising human acid glucosidase, mannitol and sucrose in aqueous solution.

60. A pharmaceutical composition prepared by lyophilizing a first composition comprising human acid alphaglucosidase, mannitol, sucrose and an aqueous solution to produce a second composition; and reconstituting the lyophilized composition in saline to produce a third composition.

61. The pharmaceutical composition of claim 60, wherein the human acid alpha-glucosidase is at 5 mg/ml in both the first and third composition, the mannitol is at 2 mg/ml in the first composition, the sucrose is at 0.5 mg/ml in the first composition, and the saline used in the reconstituting step is 0.9% w/w.

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